Creutzfeldt-Jacob Disease ("Prion" Disease)

August 2004 Updates (page 3,4)

1) THE DISEASE AND ITS EPIDEMIOLOGY

A. Etiologic Agent

Creutzfeldt-Jacob disease (CJD) is caused by an "unconventional " filterable agent, a unique protein ("prion") that replicates by a poorly understood mechanism.

B. Clinical Description and Laboratory Diagnosis

CJD has an insidious onset with confusion and forgetfulness that progress rapidly to severe cortical dementia in combination with ataxia, myoclonus and an abnormal electroencephalogram (EEG), showing typical periodic complexes.

Five prion diseases occur in humans: CJD, Gerstmann-Straussler-Scheinker syndrome, kuru, fatal familial insomnia and, described in late 1990s, a new variant of Creutzfeldt-Jacob disease (vCJD) associated with bovine spongiform encephalopathy (BSE) commonly referred to as "mad cow disease" in the popular press. CJD occurs almost exclusively (99%) in patients 35 years old and older. Characteristically, routine laboratory studies of cerebrospinal fluid (CSF) are generally negative with the exception of a mildly elevated protein level. The detection of the 14-3-3 protein in CSF has been suggested as a marker for prion disease; however false negatives and false positives associated with herpes encephalitis, hypoxic brain damage, acute stroke, and other conditions have been reported. The vCJD is notably different from that of typical CJD. It occurs primarily in younger individuals (average age 27, range 16 to 48 years) with a somewhat protracted course of approximately 16 months, and it is not associated with typical periodic complexes on the EEG. Both forms can be distinguished when biopsy or post mortem examination of the brain is performed.

Laboratory diagnosis is based on demonstration of an abnormal protein in brain tissue. The presence of the 14-3-3 protein in the spinal fluid has been reported as helpful in diagnosis.

C. Reservoirs

Human cases constitute the only known reservoir of CJD; however, there is mounting evidence supporting the idea that vCJD results from infection of humans with the agent causing BSE.

D. Modes of Transmission

The mode of transmission for most cases of CJD is unknown. In some cases transmission occurs through contaminated pharmaceutical preparations and surgical procedures (iatrogenic cases of CJD). Precautions are now in place to ensure that this does not occur. The vCJD is linked to consumption of certain parts of cattle infected with the agent causing BSE.

E. Incubation Period

The incubation period of iatrogenic CJD cases ranges from fifteen months to possibly 30 years. The incubation period of vCJD is debatable. Some researchers postulate a 5-year incubation period. Others are postulating much longer incubation period, closer to that of CJD.

F. Period of Communicability or Infectious Period

The central nervous system tissues are infectious throughout the symptomatic period of CJD cases. Other tissues and CSF are also sometimes infectious.

G. Epidemiology

CJD occurs worldwide with an incidence of roughly 1 per million populations per year. It occurs sporadically (approximately 90% of cases) through iatrogenic transmission of infective agents (<1% of cases) or as an autosomal dominant inheritance (approximately 10% of cases). Variant CJD was first described in the United

Kingdom; as of May 4, 2004, 141 cases were reported. In New Jersey, approximately 7 cases of sporadic CJD are reported every year to the NJDHSS, and no case of vCJD has been reported to date in New Jersey. One case of vCJD in the United States reported in Florida involved a person living many years in the United Kingdom.

2) REPORTING CRITERIA AND LABORATORY TESTING SERVICES

A. New Jersey Department of Health and Senior Services (NJDHSS) Case Definition

CASE CLASSIFICATION

A. CONFIRMED

A clinically compatible case supported by:

- Neuropathological examination (brain biopsy, post-mortem examination)

B. PROBABLE

A clinically compatible case supported by:

- Characteristic EEG changes (CJD), **AND/OR**
- Detection of 14-3-3 protein in spinal fluid (CJD), **OR**
- Characteristic posterior thalamic high signal on MRI scan (vCJD).

C. POSSIBLE

A clinically compatible case not supported by EEG, MRI or other laboratory findings.

B. Laboratory Testing Services Available

The Public Health and Environmental Laboratories (PHEL) does not provide testing for CJD.

3) DISEASE REPORTING AND CASE INVESTIGATION

A. Purpose of Surveillance and Reporting

- To identify disease clusters and demographic characteristics.

B. Laboratory and Healthcare Provider Reporting Requirements

N.J.A.C. 8:57-1.8 stipulates that laboratories report (by telephone, confidential fax, over the Internet using the Communicable Disease Reporting System [CDRS] or in writing) all cases of CJD to the local health officer having jurisdiction over the locality in which the patient lives, or, if unknown, to the health officer in whose jurisdiction the health care provider requesting the laboratory examination is located. The health care providers must report all cases of CJD to the local health officer having jurisdiction over the locality in which the patient lives.

C. Health Officer Reporting and Follow-Up Responsibilities.

1. **Reporting Requirements**

The New Jersey Administrative Code (N.J.A.C. 8:57-1.8) stipulates that each local health officer must report the occurrence of any case of CJD, as defined by the reporting criteria in Section 2 A above. Current requirements are that cases be reported to the NJDHSS IZDP using the Creutzfeld-Jacob Disease Reporting Form. A report can be filed electronically over the Internet using the confidential and secure CDRS. **If vCJD is suspected, immediately notify the NJDHSS IZDP at 609.588.7500.**

2. **Case Investigation**

a. It is the local health officer's responsibility to complete a Creutzfeld-Jacob Disease Reporting Form . The information required on the form can be obtained from the case-patient's healthcare provider, case-patient's family or the medical record.

b. Use the following guidelines to assist in completing the form:
   1) Accurately record the patient's demographic information, date of symptoms onset, whether hospitalized (and associated dates), outcome of disease, and whether the patient has any familial history of dementia, or history of corneal transplant. Ask if the patient lived more than 6 months in England in the last 10 years or received a blood transfusion in England since 1980. Collect information about clinical symptoms. Ask the healthcare provider to submit a copy of the medical record or enlist his/her aid in completing these sections of the case report form.
   2) **If the patient is =<55 years old or when vCJD is suspected, immediately notify NJDHSS IZDP at 609.588.7500. In these cases additional CDC forms should be completed. Forms will be supplied by IZDP.**
   3) Collect as much information as possible about electroencephalographic studies if they were done, and about biopsy or necropsy if it was done.
   4) If several attempts to obtain patient information were made (*e.g.,* the patient or healthcare provider does not return calls or respond to a letter, or the patient refuses to divulge information or is too ill to be interviewed), please fill out the form with as much information as possible. Please note on the form the reason why it could not be filled out completely.
   5) If CDRS is used to report, enter collected information into the "Comments" section.

   After completing the case report form, attach lab report(s) and mail (in an envelope marked "Confidential") to NJDHSS, or the report can be filed electronically over the Internet using the confidential and secure CDRS.
   The mailing address is:

   NJDHSS
   Division of Epidemiology, Environmental and Occupational Health
   Infectious and Zoonotic Diseases Program
   P.O.Box 369
   Trenton, NJ 08625-0369

c. Institution of disease control measures is an integral part of case investigations. It is the local health officer's responsibility to understand, and, if necessary, institute the control guidelines listed below in Section 4, "Controlling Further Spread."

4) CONTROLLING FURTHER SPREAD

A. Isolation and Quarantine Requirements (N.J.A.C. 8:57-1.10)

None.

B. Protection of Contacts of a Case

None.

C. Managing Special Situations

Reported Incidence Is Higher than Usual/Outbreak Suspected

If multiple cases of CJD occur in a city/town, or if an outbreak is suspected, investigate clustered cases. Consult with the NJDHSS IZDP at 609. 588.7500. The IZDP staff can help determine a course of action to prevent

further cases and can perform surveillance for cases that may cross several jurisdictions and therefore be difficult to identify at a local level.

Recommendations for Funeral Directors

The New Jersey Funeral Service Education has issued "Creutzfeldt-Jacob Disease a Practical Guide for the Embalmer" available **from** New Jersey Funeral Service Education Corp., PO Box L, Manasquan, New Jersey 08736; phone 800.734.3712. Guide describes precaution s which should be taken by a mortician when dealing with the body of person with CJD.

D. Preventive Measures

Transmission studies have shown that primates can be infected via percutaneous inoculation with brain and spinal cord tissues from animals and humans with CJD. CSF and other tissues are also sometimes infectious. These facts and unusual resistance of the prion to inactivation procedures necessitate special precautions in dealing with infected individuals. Tissue from an infected person can not be used in transplants. EEG electrodes and surgical instruments contaminated by tissue from such patients should be appropriately sterilized. Consumption of the meat from cattle herds infected with the agent causing BSE is banned. Blood donation should not be accepted from individuals at high risk for CJD (history of residence or travel to the UK for three month or longer during 1980—1996, and individuals traveling to other European countries for an extended period of time since 1980).

ADDITIONAL INFORMATION

A *Creutzfeldt-Jacob Disease Fact Sheet* can be obtained at the NJDHSS website at <http://www.state.nj.us/health>. Click on the "Topics A to Z" link and scroll down to the subject *Creutzfeldt-Jacob Disease*.

There is no formal Centers for Disease Control and Prevention (CDC) surveillance case definition for CJD. CDC case definitions are used by state health departments and CDC to maintain uniform standards for national reporting. For reporting a case to the NJDHSS, always refer to the criteria in Section 2 A.

REFERENCES

American Academy of Pediatrics. 2000 Red Book: Report of the Committee on Infectious Diseases, 25th Edition. Illinois, Academy of Pediatrics, 2000.

Bresnitz, E., Gerwel, M. An Evaluation of a Suspected Cluster of Creutzfeldt-Jakob Disease (CJD) in New Jersey,". 2004 http://www.state.nj.us/health/eoh/cjd2004.pdf

CDC. Probable Variant Creutzfeldt-Jacob Disease in a U.S. Resident --- Florida, 2002. MMWR. 2002;51:927-929.

Chin, J., ed., Control of Communicable Diseases Manual, 17th Edition. Washington, DC, American Public Health Association, 2000.

Holman, R.C., Khan A.S., Belay, F.D., and Schonberger, L.B., Creutzfeldt-Jacob Diseasae in the United States, 1979-1994: Using National Mortality Data to Assess the Possible Occurrence of Variant Cases. Emerging Infect Dis 1996;2:332-337.

Mandell, G., Benett J., Dolin R., Principles and Practice of Infectious Diseases. Churchill Livingstone, 2000.

Massachusetts Department of Public Health, Division of Epidemiology and Immunization. Guide to Surveillance and Reporting. Massachusetts Department of Public Health, Division of Epidemiology and Immunization, January 2001.

Mastrianni, J.A., Roos, R.P., The Prion Diseases. Sem Neurology 2000;20:337-352. Thieme Medical Publishers